Case Report

A rare case of inflammatory myofibroblastic tumour in the maxillary sinus: masquerading a malignancy

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ABSTRACT

Aim establishing the diagnosis of inflammatory myofibroblastic tumour is very challenging due to its varied clinical and radiological manifestations. We report a rare presentation of a 5 years old female child with inflammatory myofibroblastic tumour, which was endoscopically resected with postop steroid therapy. Case report, a 5 years old female child presented to ENT OPD with complaints of right nasal obstruction since, 4 months along with recurrent minor episodes of spontaneous bleeds from the right nasal cavity. On examination a huge fleshy reddish polypoidal and pulsatile mass was seen filling the entire right nasal cavity associated with right eye proptosis. MRI with contrast showed expansion of right maxillary sinus bony walls with internal soft tissue lesion measuring 41 mm (TRANS) × 36 mm (AP)×31 mm (CC) seen enhancing with contrast. Vascular supply is from maxillary artery. Under anaesthesia right external carotid artery ligation was done followed by removal of mass from right nasal cavity. Histopathological examination was consistent with inflammatory myofibroblastic tumour. On immunohistochemistry spindle cells showed cytoplasmic positivity for SMA. Inflammatory myofibroblastic tumour though a very unusual tumour, should be considered in the differential diagnoses of tumours in the nasal cavity. A vigilant clinical evaluation and extensive imaging along with histopathological examination can guide us to an early diagnosis and proper treatment resulting in better patient satisfaction.

Keywords: Inflammatory myofibroblastic tumour, Immunohistochemistry, Endoscopic sinus surgery

INTRODUCTION

Inflammatory myofibroblastic tumour has various synonyms like inflammatory pseudo tumour, plasma cell granuloma and inflammatory myofibrohistiocytic proliferation. It is a lesion characterised by spindle cells along with an array of inflammatory infiltrate of plasma cells, lymphocytes and eosinophils.1 Most common site of occurrence is lung followed by abdominal cavity, retroperitoneum and extremities. Very rare incidences have been reported in head and neck.2

Establishing the diagnosis of inflammatory myofibroblastic tumour is very challenging due to its varied clinical and radiological manifestations. It often mimics malignancy.3 Though it usually gives an indolent picture there have been cases of extensive invasion, recurrence and metastases.4 We report a strange presentation of a 5 years old female child with inflammatory myofibroblastic tumour, which was endoscopically resected with postop steroid therapy.

CASE REPORT

A 5 years old female child presented to ENT OPD with complaints of right nasal obstruction since, 4 months along with recurrent minor episodes of spontaneous bleeds from the right nasal cavity. It was also associated
with hyposmia. No other symptoms like pain, discharge, headache, ear ache and aural fullness were noted. No symptoms and signs of cranial nerve involvement. No complaints of vision loss. No previous history of trauma or any surgical procedures.

On examination a huge fleshy reddish polypoidal and pulsatile mass was seen filling the entire right nasal cavity associated with right eye proptosis. Right eye movements and vision were normal. Light reflex was present. Throat and ear examination were normal. Cervical lymphadenopathy was absent. All routine laboratory investigations were within normal limits. Non contrast computerised tomography (NCCT) imaging showed a space occupying lesion in the right maxillary sinus with mass effects over surrounding structures.

Figure 1: Magnetic resonance imaging, large ill defined T1 hypo and T2/FLAIR hyperintense lesion in right maxillary sinus with marked enhancement on contrast causing deviation of septum to left.

Magnetic resonance imaging with contrast showed expansion of right maxillary sinus bony walls with internal soft tissue lesion measuring 41 mm transverse (TRANS) × 36 mm anteroposterior (AP) × 31 mm cephalocaudal (CC) seen. Thinning of bony walls with mass effect over nasal septum causing deviation to left side and mass effect over right orbit with mild superolateral deviation of right intra orbital contents was noted. Post contrast showed enhancement with central linear non enhancing areas. The lesion was seen to get its vascular supply by right maxillary artery. No evidence of calcification.

Differential diagnoses like hemangioma, angiofibroma, sino nasal malignancy and neurogenic lesions like schwannoma were considered. Patient was planned for frozen section and further excision of mass. Under anaesthesia endoscopic biopsy was done and sent for frozen section. There was extensive bleeding pre-operative, hence packed using surgical and hemostasis achieved. Histopathological examination (HPE) of frozen section showed spindle cell lesion with inflammation - probably inflammatory myofibroblastic tumour.

After 10 days a second procedure was planned - external carotid artery ligation with excision of nasal mass. Under anaesthesia right external carotid artery ligation was done followed by removal of mass from right nasal cavity. Entire mass was removed using endoscope. The peduncle of the mass seen arising from the posterior wall of right maxillary sinus and there was associated dehiscence of lamina papyracea but orbital periosteum was found to be intact. Postoperatively patient had no orbital complications and period was uneventful.

Figure 2: Magnetic resonance imaging, mass effect over right orbit with superolateral deviation of inferomedial wall.

Figure 3: H and E sections shows benign spindle shaped cells with inflammatory cells (400X).

On gross examination, specimen was received in multiple pieces, largest measuring 3x1.5x1 cm with a firm grey white cut surface. Histopathological examination showed a tumour composed of haphazardly arranged elongated to plump spindle cells to stellate cells displaying mild nuclear pleomorphism, few with distinct nucleoli and with moderate amounts of eosinophilic cytoplasm. Mitotic activity was inconspicuous. Stroma was edematous and showed moderate inflammatory infiltrate of lymphocytes, plasma cells, histiocytes, eosinophils and
mast cells. Few cells resembling ganglion cells were also present. Morphology was consistent with inflammatory myofibroblastic tumour. On immunohistochemistry (IHC) the spindle cells showed patchy cytoplasmic positivity for smooth muscle actin (SMA) and were negative for S-100, myogenin and anaplastic lymphoma kinase (ALK).

![Figure 4: IHC, the spindle cells shows diffuse positivity for SMA (100X).](image)

Patient was started on low dose steroids, tapered over a period of 6 weeks. On follow up endoscopy of nasal cavity was clear and there was marked reduction of the proptosis. There is no recurrence till date.

**DISCUSSION**

Inflammatory myofibroblastic tumour has been first described by Brunn in 1939 as immunohistochemically diverse entity comprising both neoplastic and non-neoplastic qualities.\(^5,6\) And this term was given by Umiker and Iverson in 1954.\(^6\) This fascinating tumour has grabbed the attention of pathologist, surgeon and oncologist due its peculiar clinical behaviour, histopathological appearance and therapeutic response.\(^7\)

A variable degree of spindle cell proliferation within a background of myxoid/collagenous stroma and inflammatory infiltrate composed of lymphocytes, histiocytes, plasma cells and eosinophils are descriptive of this lesion.\(^8,9\) Most common organ of involvement is lung but can occur in multiple other sites of the body. The extra pulmonary involvement of head and neck ranges from 14-18%.\(^10\) In head and neck, larynx is the most common site.\(^11\)

In a study by Zhu et al, maxillary sinus was found to be the commonest site followed by orbit, ethmoid and nasal cavity.\(^12\) There has been no age predilection ranging from childhood to adults and equal incidences are reported in both male and female.\(^13\) Various etiologies attributed were ALK gene rearrangement, viruses like *Epstein–Barr* virus and HHV-8, trauma, chronic inflammation and autoimmune disease.\(^14\) Clinical features are mainly site specific. Nasal inflammatory myofibroblastic tumour cause nasal obstruction, bleeding and symptoms due to pressure effect over surrounding structures like proptosis in our case.

Magnetic resonance imaging and contrast enhanced computed tomography are investigations of choice to know vascularity of the tumour and extent of local infiltration due to the close proximity of vital structures like orbit. At least one sinus wall is destroyed.\(^4\) Radiological picture is often misleading to the diagnosis of malignancy due to the extensive infiltration and bony destruction.

The unpredictable immunohistochemical picture and cytopathological appearance has been categorised as 4 variants largely modified from classification by Coffin et al in 1995.\(^5,9\)

Spindle cells within a vascularized myxoid stroma and an inflammatory infiltrate of neutrophils and eosinophils. Compact spindle cells within a collagenized stroma and storiform architecture and an inflammatory infiltrate of plasma cells and lymphocytes often forming lymphoid follicles with germinal centres. Elongated spindle cells within a hypocellular highly collagenous stroma and a variable inflammatory infiltrate of lymphocytes, plasma cells, and eosinophils. Lymphohistiocytic variant consisting of myofibroblastic spindle cells and foamy histiocytes. This is thought to represent the most inflammatory variant.

There has often been an overlap among these histologic classifications.\(^9\) On Immunohistochemistry, spindle cells are reactive to vimentin (99%), smooth muscle actin (92%) and muscle specific actin (89%).\(^8\)

Similar to diagnosis treatment has also been a topic of uncertainty. Till date there are no established treatment protocols. Surgery is the first line of management along with adjuvant therapies being corticosteroids, radiotherapy and chemotherapy.\(^3\) Complete resection of the tumour results in 90% cure. Though inflammatory myofibroblastic tumour follows a favourable clinical course, there is a chance of 25% recurrence.\(^15\)

Study by He et al, showed multiple relapses, mitosis , necrosis, ganglion like cells and histological patterns were associated with poor outcomes.\(^16\) ALK status plays an important role, as it increases the chance of metastases and is more aggressive in ALK negative tumours.\(^9\) Even though this case was negative for ALK, the tumour was completely resectable and there was no evidence of metastases.

**CONCLUSION**

We would like to conclude that, inflammatory myofibroblastic tumour though a very unusual tumour, should be considered in the differential diagnoses of...
tumours in the nasal cavity. Due to the high vascular nature of tumour, proper imaging and preoperative planning is required to prevent per-operative haemorrhage. Thus, helping in complete resection of tumour. It is multidisciplinary team approach which helps in diagnosis and management of this tumour involving pathologists, radiologists and surgeons. The significance of histopathological examination and immunohistochemistry is indispensable in this tumour for establishing diagnosis and prognosis. The medical and surgical management of these patients should be individualized due to the varied clinical behaviour of the tumour. A vigilant clinical evaluation and extensive imaging along with HPE can guide us to an early diagnosis and proper treatment resulting in better patient satisfaction.

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